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CASO CLÍNICO

OHVIRA syndrome: report of a case in Bolivia

Síndrome de OHVIRA: reporte de un caso en Bolivia

Leonel Rivero Castedo[1], Jhossmar Cristians Auza-Santivañez[2], Roberto Carlos Jimenez-Fernandez[3], Elier Carrera González[4], Alba Rossio López Castillo[5], Jorge Soneira Pérez[6]

[1]Universidad Privada Franz Tamayo. Santa Cruz, Bolivia.
[2]Ministerio de Salud y Deportes. Instituto Académico Científico Quispe-Cornejo. La Paz, Bolivia.
[3]Universidad Católica Boliviana "San Pablo". Santa Cruz, Bolívia.
[4]Hospital Clínico-Quirúrgico Miguel Enríquez. Unidad de cuidados Intensivo. La Habana, Cuba.
[5]Hospital de San Lorenzo. Tarija, Bolivia.
[6]Hospital Universitario Clínico Quirúrgico "Dr. Miguel Enríquez". La Habana, Cuba.

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ABSTRACT

Introduction: the OHVIRA Syndrome was described by Herlyn Werner Wunderlich and in 1976 Wunderlich described a grouping of right renal aplasia with bicornuate uterus and simple vagina in the presence of an isolated hematocervix, as a characteristic triad uterus didelphys, intercepted hemivagina and ipsilateral renal anomaly, it is generally performed The diagnosis occurs in puberty at the beginning of menarche, with manifest symptoms of progressive dysmenorrhea and non-specific abdominal pain in the hypochondrium; urinary retention, urinary infection or a pelvic mass usually appear.
Clinical case: the case of a 13 - year - old patient is presented, who comes to the clinic with abdominal pain and transvaginal bleeding. Imaging studies are performed and due to the characteristics of said studies, the presence of OHVIRA Syndrome is suspected.
Discussion. Once the imaging studies were performed and the diagnosis confirmed, a surgical procedure was performed under general anesthesia. Through exploratory laparotomy, a bicornuate uterus was visualized, the right uterus larger than the left, and adherence to the abdominal wall, so an open intervention was decided.
Conclusions: OHVIRA syndrome coexists with a rare malformation and is often misdiagnosed as other more common etiologies of dysmenorrhea in adolescents, as a consequence it delays a correct and early diagnosis, increasing the risk of kidney damage and its complications. In our clinical case, a timely diagnosis was made and surgical treatment was subsequently planned with a favorable outcome.

KEYWORDS

Dysmenorrhea; Hematometra; Müllerian Malformations; Renal Agenesis.

RESUMEN

Introducción: el Síndrome de OHVIRA fue descrito por Herlyn Werner Wunderlich y en 1976 Wunderlich describió una agrupación de aplasia renal derecha con útero bicorne y vagina simple en presencia de un hematocérvix aislado, como triada característica útero didelfo, hemivagina interceptada y anomalía renal ipsilateral, generalmente se realiza el diagnostico en la pubertad en inicio de la menarca, con clínica manifestada de dismenorrea progresiva y dolor abdominal no específico en hipocondrio, suele aparecer retención urinaria, infección urinaria o una masa pélvica.
Caso clínico: se presenta el caso de una paciente de 13 años de edad, que acude a consulta con dolor abdominal

y sangrado transvaginal, se realiza estudios de imagen y por las características de dichos estudios se sospecha de la presencia de un Síndrome de OHVIRA.
Discusión: al realizarse los estudios de imagen y confirmar el diagnóstico se realiza procedimiento quirúrgico bajo anestesia general, mediante laparotomía exploratoria se visualiza útero bicorne, derecho mayor tamaño que el izquierdo, adherencia a pared abdominal, por lo que se decide intervención abierta.
Conclusiones: al síndrome de OHVIRA coexiste una malformación rara y a menudo se diagnostica equivocadamente como otras etiologías más comunes de dismenorrea en adolescentes, como consecuencia retrasa un diagnóstico correcto y temprano, aumentado el riesgo de daño renal y sus complicaciones. En nuestro caso clínico, se dio un diagnóstico oportuno y posteriormente se planificó un tratamiento quirúrgico con una evolución favorable.

PALABRAS CLAVES

Dismenorrea; Hematómetra; Malformaciones Mullerianas; Agenesia Renal.

INTRODUCTION

OHVIRA syndrome was described in 1925 by Herlyn Werner Wunderlich, [1] according to Zhu et al. He relates that it was initially described in 1971 by Herlyn-Werner, [2] and in 1976 Wunderlich described a grouping of right renal aplasia with the bicornuate uterus and simple vagina in the presence of an isolated hematocrit [3] as a characteristic triad didelphic uterus, intercepted hemivagina and ipsilateral renal anomaly, the diagnosis is usually made at puberty at the onset of menarche, with clinical manifestations of progressive dysmenorrhea and non-specific abdominal pain in the hypochondrium, urinary retention, urinary tract infection or a pelvic mass. The incidence of this pathology is estimated at 0.1 to 3,8 %, [4] representing 0,16-10 % of all mullerian malformations (occurring in 1/2,000 to 1/28. 000 women), the etiology is usually unknown, but it can be originated by the unprecedented development of the Müllerian and Wolffian ducts,[5] around the eighth week of gestation,[6] the Müllerian or paramesonephric ducts fuse forming the uterus, the proximal two thirds of the female organ and the cervix with a central septum, which begins to be reabsorbed at nine weeks and finally leaves a single cavity, depending on the height of the defect, the duplicity in the HWWS ends at the uterine or cervical level, or reaches lower, having a duplex vagina, the incidence of ipsilateral anomaly is 100 %, this is due to the interaction relationship between the paramesonephric and mesonephric ducts during renal development, [6] it is a challenge the early diagnosis, besides it is not usually a single presentation, renal agenesis can be accompanied by other ipsilateral renal anomalies, [7] ectopic ureter, ureterocele and vesicoureteral reflux (VUR), contralateral urological anomalies such as VUR, mega ureter and dysplastic kidney, [6] in addition to clinical hematocolpos, irregular menstruation, such as severe dysmenorrhea or amenorrhea. Another variant is cervical atresia and a double cervix with hemicervical obstruction with a rudimentary hemiuterus; in a large percentage of OHVIRA syndrome, the septum is thick and limits the distension of the hemivagina, resulting in retrograde bleeding causing endometriosis. [7]
In patients with OHVIRA syndrome, ovarian function is not compromised, and reproductive condition is affected by problems such as endometriosis, obstructed hemiuterus pregnancies, development of pelvic abscesses, and abdominal wall adhesions have been described.
We present the case of a 13-year-old female patient who comes to the clinic with abdominal pain and transvaginal bleeding. Imaging studies are performed, and due to the characteristics of these studies, the presence of OHVIRA syndrome is suspected.

CLINICAL CASE

A female patient of 13 years of age comes to consultation with a clinical picture of abdominal pain and transvaginal bleeding to the anamnesis refers to not remembering the date of last menstruation FUM (irregular menstruation), physical examination at the abdominal level is evident infra umbilical middle scar and drainage points in surgical points Mc Burney, this as background one year ago of an exploratory laparotomy, also refers intervention with uterine drainage, aspiration for biopsy of both uterus and permeabilization of the cervix. Gestations: 0, Abortions: 0, Births: 0, flat abdomen, positive hydrothermal sounds (+), painful to superficial and deep palpation in hypogastrium, both iliac fossae, normal external genitalia, no evidence of transvaginal bleeding and no cervix palpation; with negative pack test, normal hemogram, normal blood chemistry and normal cuagulogram, fibrinogen 310mg/dL (200 - 400 mg/dL), factor O Rh+, negative CRP, normal general urine test, negative urine culture.
A gynecological ultrasound was performed to be compatible with hematometra (Vol. 291,44). Abdominal ultrasound showed two solid masses in the pelvic excavation area, one measuring (7,6 x 5,8 x 5,8 x 5,6 cm) and

the other (8,7 x 5,9 x 5,5 cm), both joined in a flange, with no endometrial image observed, the right kidney was absent, left kidney measured 11,2 x 8,2 cm, slight pyelocaliceal dilatation is observe (Figure 1A- 1B).

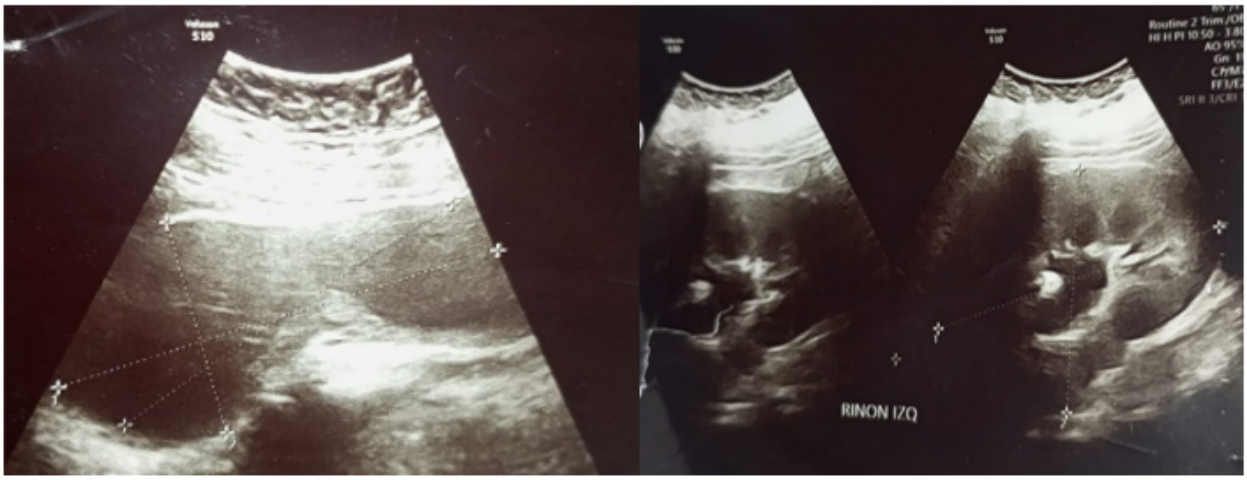

Figura 1. A: Ecografía ginecológica, evidencia imagen econegaticas compatible con hematómetra, dos masas sólidas, ambas se unen por una brida, miometrio homogéneo, útero en anteroversoflexión, útero didelfo, saco de Douglas libre. **B:** Ecografía abdominal, que evidencia presencia de riñón izquierdo y dilatación de los calices menores en forma moderada, agenesia renal derecha

In the abdominal and pelvis tomography without contrast, the left kidney was observed at the abdominal level, right renal agenesis, adjacent to it, there was a nodular image of 14 mm, with a spleen attenuation coefficient of 42UH; at the pelvis level, there was a preserved bladder, a didelphic uterus associated with hematometra, the other organs were preserved. In addition, urotomography is performed where the previous conclusion is confirmed, obtaining the measures of the left kidney of 12,8 cm longitudinal, 72 cm transverse, 69 cm anteroposterior, and 69 cm anteroposterior.

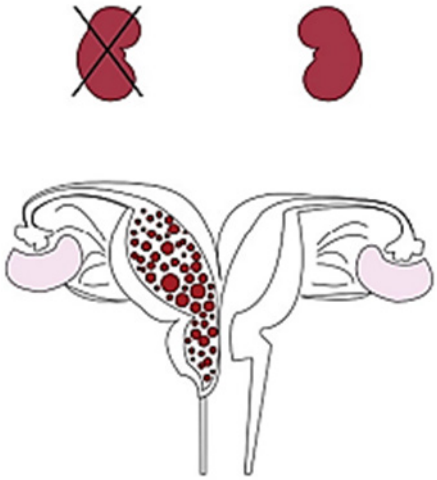

Figure 2. Graphic representation of the triad of OHVIRA syndrome (hemivaginal obstruction and ipsilateral renal impairment), of the reported case

Medical conduct

After performing imaging studies and confirming the diagnosis, a surgical procedure was performed under general anesthesia by exploratory laparotomy; a bicornuate uterus was visualized, the right larger than the left (+/- 4 cm), adherence to the abdominal wall, so it was decided to perform open surgery. The urology service team intervened with cystoscopy and placement of a double "J" catheter in the left kidney, with fluoroscopy control, the catheter was in good position, and a Foley catheter N°16 Fr was placed; Then an infraumbilical median incision is made with excision of anterior scarring, in the abdominal cavity multiple adhesions are visualized to the anterior wall and anterior face of the uterus, visualizing two uteri, the right one of greater size, with multiple adhesions that

make it impossible to see the fallopian tubes, if the right ovary is visualized, the left uterus of smaller size (+/- 5 x 4cm) which is attached at the bottom of the larger uterus, with free and visible tubes and ovaries, vertical hysterotomy is performed (+/- 6cm), entering the uterine cavity, obtaining dense liquid, dark color, not fetid, which is aspirated and digital exploration is performed, where there is no evidence of communication to the left uterine sketch, but no duct was excavated towards the cervical orifice, with obstruction in the lower part of the cervix, in the exploration by vaginal canal, suprapubic valva is placed, channeling and forming new cervix and communication with the uterine cavity, then endotracheal tube N°8 is introduced into the uterine cavity, through the cervical canal, passing through the vaginal canal until it reaches the external part, where it is fixed on the inner side of the left thigh, all adhesions are removed, hysterorrhaphy is performed, empirical broad-spectrum antibiotic therapy is indicated, the management of this patient was multidisciplinary.

DISCUSSION

The classification of OHVIRA syndrome is by clinical features in complete or incomplete obstruction of the hemivagina, as follows: Classification 1, patients with complete obstruction of the hemivagina, and classification 2, patients with partially obstructed hemivagina. In the follow-up of the patients in a period of 1 to 120 months, the median was 17 months; renal agenesis favored the right side, all patients underwent resection of the vaginal septum and drainage of the hematocolpos, some women were married and sexually active, 85 % of the women who decided to conceive became pregnant, the pregnancy occurred in a higher percentage in the uterus contralateral to the hemivaginal septum, other women experienced separate pregnancies in each of the bilateral uteri, in this group there were no pathological pregnancies or pregnancy complications.[2]
The classification of Mullerian anomalies, according to Velandia et al., had improved since 1979 when they were described by Buttiram and Gibbonsen, the AFS (American et al.) published a classification that was used for decades, and in 2004, Acien and co-authors generated a system based on the embryological origin of the malformations, considering the pathophysiology and the various related organs in the development of the female genital system, different from them Oppelt P and participants describe the VCUAAM system (Vagina et al. Associated Malformation), integrating findings found in the vagina, cervix, uterus, and adnexa, and the associated anomalies were assigned to a group M, relative to each specific group. However, it was observed that several congenital malformations were not contained in the important categories or subcategories of this system.
For example, the cervical septate uterus with or without a vaginal septum, the didelphic uterus with an obstructive vaginal septum, and the bicornuate uterus with cervical or vaginal aplasia. This is why the European Society of Human Reproduction and Embryology (ESHRE) and the European Society of Gynaecological Endoscopy (ESGE) developed a new system established under the name CONUTA (Congenital et al.), which is based on the morphology of the female genital tract, embryological origin as the main class, cervical and vaginal anomalies are classified in independent subclasses, with clinical disposition being the main initial site to make a good diagnosis and to establish treatment with alternatives, however the arcuate uterus is excluded from this classification.

- Using the tables to make the diagnosis, the challenge of which method to use continues, based on ultrasound, invasive methods, and others based on high-quality images, sensitivity, and specificity; their contraindications and limitations for each patient must be taken into account.

The determination of OHVIRA syndrome is mainly based on the following complementary examinations:

- Pelvic ultrasound: It is the first line of diagnostic imaging. It allows for identifying uterine anomalies, detecting partial vaginal obstruction by the vaginal septum, and evaluating the kidneys. It has a sensitivity of 78-90 % for OHVIRA, with a 92,1 % of use. [8]
- Renal ultrasound: Evaluates the presence or absence of ipsilateral kidney. It is useful since 50 % of OHVIRA is associated with renal agenesis.
- Magnetic resonance imaging (MRI): Gold standard for soft tissue resolution. It may be evidence of the obstructive origin of hematocolpos. Sensitivity >95 %. With 74,6 % of use. [8]
- Voiding cystourethrography is the gold standard for ruling out vesicoureteral reflux of the ipsilateral kidney.
- Urotomography provides detailed 3D images of the urinary tract, with lower sensitivity to detect low-grade reflux, and is usually more expensive, with less exposure to radiation.
- Laparoscopy: In ambiguous cases, it allows for the visualization and corroboration of uterine and tubal anomalies.
- Biopsy: Histological analysis of the vaginal septum to rule out other causes of obstruction.

There are risks before surgery of making an incomplete incision and presenting the need for a new surgery, in addition to predisposing to endometriosis and complications in case of pregnancy that may occur in patients

with OHVIRA syndrome previously corrected surgically include Spontaneous abortion, preterm labor (associated with increased uterine contractions and premature cervical changes, the rate can be as high as 50 %), Restricted intrauterine growth (due to uterine vascular anomalies leading to placental insufficiency), abnormal fetal presentation, preeclampsia, postpartum hemorrhage (due to uterine hypotonia or placental retention due to anatomical abnormalities), infertility (due to recurrent synechiae or tubal blockages after corrective surgery), uterine rupture (very rare, but possible due to weakness of the muscular wall), renal complications (impaired renal function in pregnancy), it is recommended not to consume NSAIDs. Preconception counseling and close prenatal controls are recommended in the future, being essential for the early detection and timely management of these potential obstetric complications.[7] According to Candiani et al. [9], more than 85 % of the patients who sought pregnancy were successful, as is the case of Reis et al. [10]

Similar clinical cases were reported in other countries, as in the case of Kueppers et al. [6] using the VCUAM classification and the specific classification elaborated by Zhu et al., with one case being "Classification 1" "Subclass 1. 1", due to acute clinical presentation, avoiding complications that can evolve rapidly; with 'Classification 2' there is a post-puberty clinical presentation, presenting years after menarche. [6] We must consider differential diagnoses in neonates, such as embryonic rhabdomyosarcoma (botryoid sarcoma), ureteral prolapse, hymenal or vaginal cysts (Gartner), prolapsed ectopic ureterocele, hydometrocolpos.[6,11]

It is important to avoid complications due to hemivaginal obstruction, which is an indication for surgery and drainage of the hydrocolloids after the operation, to monitor for possible recurrent obstruction of the hemivagina due to renal anomalies, particularly in patients under five years of age. [6] Chan et al. report the case of fetal autopsy, giving OHVIRA results, which was performed on the fetus born dead in the third trimester, revealed a didelphic uterus, obstructed left hemivagina and a left pelvic atrophic duplex kidney, with left ureters entering obstructed left hemivagina, associated with anorectal malformation, a single right umbilical artery and spina bifida occulta. [12] The literature also describes OHVIRA syndrome in its late presentation in a 14-year-old adolescent with a rare variant of this syndrome; the authors highlight the relatively long delay in diagnosis two years after menarche, attributed to the presence of a contralateral non-obstructive hemivagina that allowed a partial menstrual flow,[9,13] which in some cases the relative management and Antibiotic therapy.[14] The diagnosis was made by abdominal ultrasound and confirmed by abdominal and pelvic tomography and urotomography. The medical treatment administered in the case was micronized progesterone 200 mg daily dose case scheduled surgery, as in most cases.[2] As we can see, the conduct has been the same in most reports; the diagnosis was by ultrasonography, urotomography, and magnetic resonance imaging, being the gold standard, and the definitive management was corrective surgery.

CONCLUSIONS

OHVIRA syndrome coexists with a rare malformation and is often misdiagnosed as other more common etiologies of dysmenorrhea in adolescents; as a consequence, it delays a correct and early diagnosis, increasing the risk of renal damage and its complications.

In our clinical case, a timely diagnosis was made, and subsequently, surgical treatment was planned with a favorable evolution. We consider that knowledge of the pathophysiological characteristics, clinical presentation, and diagnosis is the best tool for the timely medical-surgical therapeutic management of these patients and to avoid their suffering from complications.

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CONSENT

The patient's consent was obtained for this work.

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CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

AUTHORSHIP CONTRIBUTION

Conceptualization: Leonel Rivero Castedo.
Research: Jhossmar Cristians Auza-Santiváñez, Leonel Rivero Castedo.
Methodology: Jhossmar Cristians Auza-Santiváñez, Leonel Rivero Castedo.
Visualization: Roberto Carlos Jiménez Fernández, Jorge Soneira Pérez, Elier Carrera González.

Original draft: Leonel Rivero Castedo, Jhossmar Cristians Auza-Santivañez, Roberto Carlos Jimenez-Fernandez, Elier Carrera González, Alba Rossio López Castillo, Jorge Soneira Pérez.
Writing-revising and editing: Leonel Rivero Castedo, Jhossmar Cristians Auza-Santivañez, Roberto Carlos Jimenez-Fernandez, Elier Carrera González, Alba Rossio López Castillo, Jorge Soneira Pérez.